Left main coronary artery atresia masquerading as dilated cardiomyopathy treated with aortic reimplantation

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Left main coronary artery atresia (LMCAA) is rare, and only 19 pediatric patients with this lesion have been reported in the literature.1, 2 Early diagnosis of LMCAA with aggressive surgical treatment is usually lifesaving. This report describes the case of a 7-year-old girl with LMCAA with heart failure that had been masquerading as dilated cardiomyopathy since infancy. The symptoms and signs completely resolved after coronary reimplantation.

Clinical Summary

A 7-year-old girl was seen with dyspnea and shortness of breath at the age of 4 months. A diagnosis of dilated cardiomyopathy with severe mitral regurgitation was made after initial echocardiographic study. After this diagnosis, she was treated with digoxin, furosemide, captopril, and aspirin, with stable condition. Follow-up echocardiography showed a dilated right coronary artery, but the ostium of the left main coronary artery was difficult to delineate. In addition, a reversed flow in the left coronary artery was demonstrated on color Doppler scan.

Cardiac catheterization revealed the pulmonary to systemic flow ratio at the pulmonary artery level to be 1.0. The left ventriculogram demonstrated a dilated and impaired left ventricle (ejection fraction 0.42), with moderate mitral regurgitation. The left coronary artery could not be selectively catheterized at either the aortic root or the pulmonary artery root. Right coronary angiography showed a dilated right coronary artery, with abundant collateral vessels draining to the left coronary artery. The patient was operated on for presumed LMCAA with moderate mitral regurgitation.

At operation, both great arteries were transected, and the absence of the left main coronary orifice in both aortic and pulmonary artery root was confirmed. The proximal left coronary artery was found to be intramural and blind ended (Figure 1). A diagnosis of LMCAA was established, and the left main coronary artery was subsequently reimplanted to the aortic root.

Three years after the operation, follow-up cardiac catheterization revealed a dramatic improvement in left ventricular function (ejection fraction 0.60), with significant regression of mitral regurgitation and right coronary dilation. Left coronary angiography revealed a widely patent reimplanted coronary artery (Figure 2).

Discussion

Most patients with LMCAA have symptoms, with failure to thrive and myocardial infarction during infancy and frequently syncope and tachyarrhythmias during childhood and adolescence. On extremely rare occasions, LMCAA may be asymptomatic.1, 3 Includ-
ing this report, only 3 cases of LMCAA have been described as manifesting as dilated cardiomyopathy.\textsuperscript{4,5} If cardiomyopathy is caused by congenital coronary anomalies, its recognition and prompt surgical treatment radically modifies the prognosis. coronary artery bypass grafting seems to be the procedure of choice for adult patients, but the long-term results of bypass grafting in pediatric patients are questionable, although good results have been reported.\textsuperscript{1-5} Direct surgical reimplantation or reconstruction of the left main coronary artery offers several advantages relative to bypass grafting. Occlusion of the main trunk is prevented, antegrade flow is provided, competitive flow is avoided, bypass material is spared, percutaneous transluminal angioplasty may still be performed, reoperative surgery can be easily performed, and the shortest and most efficient way for blood to the myocardium is provided. However, application of this elegant technique for LMCAA is not common and has been reported in only 1 case.\textsuperscript{3} We therefore advocate reconstructing the left main trunk with direct reimplantation, as in our case, or with a baffle of ascending aorta as the procedure of choice for treating this particular coronary anomaly whenever feasible.

The management of the ischemic mitral regurgitation at the time of congenital coronary artery anomalies repair is controversial. There are only two reports regarding mitral surgery addressed concomitantly at the time of LMCAA repair.\textsuperscript{2,3} We believe that if the mechanism of severe mitral regurgitation in the older child or adolescent is caused by irreversible myocardial injury or papillary muscle infarction, then repair of the mitral valve at the time of coronary reimplantation may be warranted. In contrast, if the patient is without any irreversible myocardial damage, as in our case, associated mitral regurgitation may improve after repair as myocardial ischemia, ventricular dilation, and papillary muscle dysfunction improve.

In summary, this particular case illustrates that LMCAA may masquerade as dilated cardiomyopathy with severe mitral regurgitation. The recognition and prompt surgical treatment of this lesion, especially with anatomic repair by aortic reimplantation, radically modifies the prognosis. Improvement in mitral regurgitation, normalization of ejection fraction, and lessening of left ventricular dilatation are anticipated.

References